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An Alternative Treatment for Type B Ulnar Polydactyly



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An Alternative Treatment for Type B Ulnar Polydactyly

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Based on a study performed at Red Cross Children's Hospital, presented at the South African Society for Surgery of the Hand in August 2006 and August 2008.

Declaration

I, **Michelle Nerine Maree**, hereby declare that the work on which this dissertation is based is my original work (except where acknowledgements indicate otherwise) and that neither the whole work nor any part of it has been, is being, or is to be submitted for another degree in this or any other university.

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Summary

Rudimentary ulnar polydactyly is one of the most common congenital hand anomalies. These are conventionally treated by suture ligation in the neonatal period or by formal excision, when the child is one year of age. For the last three years, the Congenital Hand Unit at Red Cross Children's Hospital has used vascular clip ligation as an alternative method of treatment for rudimentary ulnar polydactyly, based on the same principle as suture ligation, but with less associated complications.

A study was performed at the unit, where two hundred and nineteen supernumerary digits were treated over a two-year period. The digits were ligated using a vascular clip, as an outpatient procedure.

At one week, the stump had necrosed leaving no residual nubbin. In only two of the 219 digits, the clips had fallen off prematurely. No complications were noted at three-month follow-up.

In this study, vascular clip ligation of rudimentary ulnar polydactyly is a better alternative to conventional treatment. It is more cost-effective than formal excision, complications of anaesthesia and surgery are avoided, as well as complications associated with suture ligation, such as knot slippage, residual nubbin, venous engorgement and sepsis.

Introduction

Extra digits are found in all animal species. In humans, they are found across all cultural and geographical backgrounds.¹

Post-axial or ulnar polydactyly is one of the most common congenital hand anomalies with an incidence of one in 300 live births.^{1,2,3}

The simplest and most commonly used classification of ulnar polydactyly is that by Temtamy and McKusick,^{4,13} where Type A is a fully developed extra digit which articulates with the fifth metacarpal, and Type B is rudimentary and pedunculated.^{4,5,6}

Type B polydactyly is conventionally treated either by suture ligation in the immediate post-natal period, or, by formal excision under general anaesthesia when the child is approximately one year of age. However, these treatment modalities are not without significant reported complications.^{5,7,8,9}

An alternative method of treatment is being used at Red Cross Children's Hospital Congenital Hand Unit, based on the same principle as suture ligation, but using a vascular clip instead, with excellent results and minimal complications at 3-month follow-up.

Classification

There are several classification systems in use, highlighting the controversy surrounding not only the classification of ulnar polydactyly but congenital hand anomalies in general. Most classification systems are based on morphology alone and do not indicate the embryological basis for the defect, the site of the insult to the limb bud, nor the timing of such an insult.¹² Many do not take into account the tremendous variety and spectrum of expression within one group.

Many of these classification systems also provide no guidelines with regard to management decisions. A descriptive classification which would take into account, site, size, tissue elements, level, stability, mobility and the extent to which the anomaly influences the function and appearance of the hand, would be ideal, but obviously this does not lend itself to a usable classification system.¹

Despite this, the most satisfactory and widely accepted classification system in use is still that by Swanson, adopted by the International Federation of Societies for Surgery of the Hand (IFSSH) (Table 1). The aim of this classification system is to use simple terms to describe the abnormality and avoid complex Greek or Latin terms, which may be misunderstood or misinterpreted. Again, the classification implies nothing regarding the cause of the anomaly.⁶

Table 1: Swanson / IFSSH classification

- | |
|--|
| <ul style="list-style-type: none">I - Failure of formation of parts (arrest of development)II - Failure of differentiation (separation of parts)III - DuplicationIV - OvergrowthV - Undergrowth (hypoplasia)VI - Congenital constriction band syndromeVII - Generalised skeletal abnormalities |
|--|

Ulnar or post-axial polydactyly falls within the subgroup, Duplications, according to Swanson's classification of congenital hand disorders. Currently, polydactyly is divided into 3 categories: **ulnar** or post-axial (duplication of the fifth digit), **central** (duplication of the second, third or fourth digit) and **radial** or pre-axial polydactyly (duplication of the thumb).^{10,11}

There are various classification systems used to describe ulnar polydactyly. The simplest and most commonly used classification is that by **Temtamy and McKusick**:^{4,13}

Type A: the extra digit is fully developed and articulates with the fifth or an extra metacarpal.

Type B: the extra digit is rudimentary and pedunculated.^{3,5,6,13} This classification system is based on genetics, after the authors noted that parents with Type A polydactyly could have offspring with either Type A or B polydactyly, whereas parents with Type B could only produce offspring with Type B polydactyly.^{13,14}

Many have argued that this classification is too simplistic, as it ignores the wide spectrum of expression of the extra digit and does not take into account the degree of bifurcation of the ray.^{14,15} It fails to assist in

surgical planning, because it does not impart enough clinically relevant information regarding the digit.^{1,14,15}

Stelling¹⁶ and Turek¹⁷ also classified ulnar polydactyly into three types (Table 2) based on management, but this classification system is not widely used because it did not describe the pedunculated type, the most common type, adequately.¹⁴

Table 2: Stelling¹⁶ and Turek¹⁷ Classification

<u>Type</u>	<u>Morphology</u>	<u>Management</u>
Type 1	Duplication of soft tissues only	Ligation or excision
Type 2	Partial duplication, including bony structures	Simple disarticulation
Type 3	Complete duplication of the ray, including the metacarpal (<i>very rare</i>)	Formal ray excision

Rayan and Frey³ expanded on Stelling's classification, proposing a more detailed classification of ulnar polydactyly into five sub-types, based on the recognition that there is a wide spectrum of presentation within this group.

Table 3: Rayan and Frey³ Classification (Modified by Al-Qattan¹⁴)

<u>Type</u>	<u>Morphology</u>	<u>Management</u>
Type I	Simple, wart-like nubbin	No treatment
Type II	Pedunculated non-functioning digit	Ligation / Excision
*Type II A	Less than 3mm pedicle	
*Type II B	Wider than 3mm pedicle	
Type III	Well-formed digit articulating with the fifth metacarpal	Surgical excision
*Type III A	Bifid metacarpal head	
*Type III B	Proximal phalanx fused to the fifth metacarpal	
Type IV	Fully developed sixth digit with its own metacarpal shaft	Surgical excision
Type V	Associated with syndactyly	Surgical reconstruction

**As modified by Al-Qattan¹⁴*

Al-Qattan¹⁴ modified Rayan and Frey's³ classification by dividing Types II and III into 2 subtypes, a narrow (Type IIA) and wide (Type IIB) digit. The distinction in Type II was emphasised because the narrow pedicle of Type IIA was at higher risk of becoming strangulated, causing oedema and vascular compromise. Thus, earlier treatment was advised. In Type IIIA, surgery is more complicated than for Type IIIB, as the former requires collateral ligament reconstruction.¹⁴

Blauth and Olason¹⁵ have devised a complicated classification system, which they feel is appropriate to all forms of polydactyly, including that of duplication of the feet. Essentially the classification considers alterations in both the transverse and longitudinal planes of the hand. The longitudinal structure of the ray constitutes the basic framework for the classification and is classified according to the level at which the bifurcation of the digit occurs. For example, a *proximal phalanx type* malformation is when all the phalanges of the digit are abnormal i.e. the distal and middle phalanges are completely duplicated, but the proximal phalanx may be either partially or completely bifurcated up to the metacarpophalangeal joint. Special provision is made for the rudimentary types of polydactyly within the longitudinal classification, and here it is classified according to the level at which the rudimentary digit branches off. Thus a supernumerary digit attached at the level of the proximal phalanx will be ascribed the designation *rudimentary form of a proximal phalanx type*.¹⁵ The second leg of the classification is the transverse structure of the limb. Each digit is assigned a number, the thumb (or big toe), I, index finger II and so on. If the polydactyly involves two or more digits, then all the affected rays are assigned with the appropriate number.¹⁵ The classification becomes even more complicated when considering multiple duplications and will not be discussed further. This classification system is logical, however is not in common clinical use, probably because it is not easy to use or remember.

It is almost identical to the classification described earlier by **Buck-Gramcko and Behrens**¹⁸ and recommended by Light.¹⁹ This classification also has two parts: a transverse axis, allocating a number to the digit that is duplicated, and two, a longitudinal axis from distal to proximal, indicating the bony level of duplication as well as to differentiate the rays into ten types according to their duplication, assessed both anatomically and radiologically.¹⁸ For example, ulnar polydactyly duplicating at the level of the proximal interphalangeal joint would be assigned the type, *Type V PIP*.^{14,19}

Despite the general lack of consensus regarding nomenclature and classification of ulnar polydactyly, for further purposes however, reference will be made to the classification by that of Temtamy and McKusick.^{4,13}

Incidence

The true prevalence of ulnar polydactyly is unknown, as the majority of these are seen in the neonatal unit and tied off by paediatricians or obstetricians without being reported.^{1,5,20,21}

Temtamy and McKusick Type B is the most common abnormality in a population of African descent, occurring in approximately one in 300 live births. In South Africa, however, it is even more common with an incidence of greater than one per 100 live births.^{1,2} In the Caucasian population, the incidence is quoted as one in 1500 to 3000 live births.^{1,3,6,10,14,20,21}

Type A on the other hand is very rare, and occurs in approximately 0.014% of all live births.^{10,22} It occurs with the same frequency amongst both the African and Caucasian populations.¹⁰

In African and Caucasian populations, ulnar polydactyly is more common in males than females (ratio of 1.6:1),³ but is more common in females (ratio: 1.3:1) in a Saudi Arabian population according to a study done by Al –Qattan.¹⁴ There is a frequency of eight to one when compared to the duplication of other digits.^{1,5,20} Ulnar polydactyly is more common than radial polydactyly in the African population, whereas the opposite holds true for Caucasian and Asian populations.^{5,10}

Inheritance

Types A and B are two different and independent genetic entities with different aetiological mechanisms.¹⁰ The genes involved are from two different loci.^{10, 14, 22} Though both are regulated by an autosomal dominant pattern of expression,²³ Type A also appears to be regulated by genetic modifiers, and some studies suggest autosomal recessive patterns as well.^{4,8,10}

Both Types A and B ulnar polydactyly may be either isolated or syndromic.^{3,20,24} When it occurs as an isolated clinical finding, it is usually inherited in an autosomal dominant mode, whereas when it is associated with a syndrome, it is usually inherited in an autosomal recessive mode.^{5,10, 20} Penetrance is variable, most often incomplete, and expressivity is variable.²¹

An African child presenting with a type B anomaly is usually normal with no other associated abnormalities. They more commonly have bilateral involvement, but when unilateral, there is a left sided preponderance.^{3,10,14,22} The reason for this has not been fully established.

However, when Type B ulnar polydactyly presents in a Caucasian child, it is often unilateral and associated with an underlying syndrome in 10% of cases.^{1,3,6,11,20,25} There are more than 50 described syndromes associated with ulnar polydactyly (See Table 5).^{3,10, 21} The most common examples are Trisomy 13 and Laurence-Moon-Bardet-Biedl syndrome, the latter characterised by ulnar polydactyly, pigmentary retinopathy, obesity, polycystic kidneys, hypogonadism and mental retardation.^{1,3,24,26} Other more rare syndromes are Meckel-Gruber syndrome, characterised by the triad of ulnar polydactyly, meningoencephalocoele and polycystic kidneys, as well as Ellis-van Creveld syndrome, characterised by findings of ulnar polydactyly,

disproportionate dwarfism, ectodermal dysplasia and heart defects.¹¹ Interestingly, there is a negative association between ulnar polydactyly and Down syndrome.²⁷

There are also a few non-syndromic associations, the commonest being syndactyly (51%), and polydactyly of the feet,²⁷ but more serious associations can involve any organ system. Examples of such associations include an imperforate anus, polycystic kidneys, renal agenesis, hydrocephalus, cardiac defects, co-arcuation of the aorta and clubfeet.^{10,14,27}

Therefore, though it may not be clinically obvious, ulnar polydactyly in a Caucasian patient warrants referral to a paediatrician or preferably a geneticist, and further investigation, especially of the cardiac and renal systems, is warranted in this patient group.^{1,3}

Furthermore, Zimmer and Bronshtein²⁴ suggest that the association between ulnar polydactyly and an associated syndrome or congenital abnormality is so strong, that the presence of ulnar polydactyly during prenatal ultrasound screening be used as a marker for possible congenital abnormality. In their study, 18155 fetuses were screened via vaginal ultrasound, and ten weeks gestation was the earliest a supernumerary digit could be identified. Twenty-six cases of polydactyly were detected, of which ten had associated structural abnormalities found at the same scan. In sixteen cases, the polydactyly was the only finding, and amniocentesis confirmed a normal karyotype in all cases. Post-natal follow-up confirmed that there were no false positives, however, they could not confirm their false negative rate. Thus, they recommended a detailed examination of all organ systems where ulnar polydactyly is detected and that patients should be offered amniocentesis where there is no family history of ulnar polydactyly.²⁴

Table 4: The two clinically described types of Type B polydactyly^{5,14}

<u>Clinical features</u>	<u>African Child</u>	<u>Caucasian child</u>
Incidence	1 in 300	1 in 1500 - 3000
Positive Family history	11 - 90% ⁵	5%
Limbs involved	Bilateral (70%)	Unilateral (80%)
Type of polydactyly more common	Post-axial	Pre-axial
Associated non-syndromic abnormalities	Rare 1% ¹⁴	30% ¹⁴
Associated syndrome	Rare 1% ¹⁴	10% ¹⁴

Table 5: Associated syndromes^{3,19}

Trisomy 13
Trisomy 18
Pseudotrismy 13
Laurence-Moon-Biedl-Bardet syndrome
Biedmont type 2
McKusick Kauffman syndrome
Infantile thoracic dystrophy of Jeune
Achondroplasia (Greib type)
Golabi-Rosen syndrome
Pallister-Hall syndrome
Smith Lemli Opitz syndrome
Curry Hall Syndrome
Reese Syndrome
Bloom Syndrome
Acrodermal dysostosis
Greig syndrome
Acrocallosal syndrome
Ellis van Creveld syndrome
Meckel syndrome
Carpenter syndrome
Corrodi syndrome
Goltz syndrome
Gorlin syndrome
Short rib-polydactyly syndrome
Ullrich- Frichtiger syndrome
Oral-facial-digital syndromes
Oto-palato-digital syndrome Type 2

Embryology

In normal embryonic development of the hands and digits, the limb bud develops around day 26, the hand paddles appear on day 33 and the rays appear on day 41.¹⁰ Initiation of limb bud development can be seen when the underlying mesoderm forms an outgrowth into the overlying ectoderm.^{6,28} The mesoderm is derived from two sources, the somites and the lateral plate.^{6,28} The somatic mesoderm will eventually form the muscles of the limb and the lateral plate develops into bone and cartilage.^{6,28} Between days 37 and 47, the hand paddle transforms into digital rays by digital separation, through the process of physiological cell death (apoptosis) of the interdigital cells, accompanied by active epithelial invagination.^{1,6,28,29} This process is complete within 7 days.^{1,10,28,30} By day 60, all the limb structures are present.⁶

Growth of the limb bud occurs in a controlled manner and on three spatial axes (Table 6):

1: Proximo-distal axis: this defines the outgrowth of the limb from a proximal to distal direction, and is under the control of the apical ectodermal ridge (AER) which is present in the distal end of the limb bud.^{6,28,30} The AER is a ridge of ectoderm where the volar and dorsal layers of ectoderm meet. Fibroblast Growth Factors are the primary modulators of growth within the AER.²⁸

2: Anteroposterior axis: this defines the radio-ulnar growth and is controlled by the Zone of Polarising Activity (ZPA) which is present under the ulnar border of the AER. The Sonic Hedgehog (Shh) protein is secreted by cells within the ZPA and is the primary modulator of growth within the ZPA.^{6,28,30} Shh induces Fibroblast Growth Factor 4 in the AER, providing a positive feedback loop between the AER and ZPA. Thus, there is a complex interaction between the AER and the underlying mesoderm, ensuring proportionate growth of the limb bud.²⁸

3: Dorso ventral axis: this defines formation of the dorsal and palmar structures of the hand and is controlled by the dorsal and ventral ectoderms as well as the dorsal mesoderm.⁶ The wingless type mouse mammary tumour virus integration site family member 7a gene (Wnt 7a) in the dorsal ectoderm; engrailed-1 gene (En1) in the ventral ectoderm; and LIM Homeobox transcription factor (Lmx-1) in the dorsal mesoderm modulate growth here.³⁰

The molecular pathways along all three axes are interdependent and constantly interact to ensure co-ordinated growth of the limb bud.^{28,30}

Table 6: Three spatial axes of limb development³⁰

<u>Axis</u>	<u>Location</u>	<u>1° Growth Factors</u>
Proximo-distal	Apical Ectodermal Ridge (AER)	Fibroblast growth factors
Antero-posterior	Zone of polarising activity (ZPA)	Sonic Hedgehog protein (Shh)
Dorso-ventral	Dorsal and ventral ectoderm; dorsal mesoderm	Wnt 7a; En-1; Lmx-1

Prolonged interaction between the apical ectodermal ridge (AER) and mesoderm leads to delayed involution¹⁰ and inappropriate stimulation of these mesodermal elements past the stage of apoptosis may lead to polydactyly.¹ The causes of polydactyly on a biological and molecular level are complex, multifactorial and not entirely clear. Sonic Hedgehog protein (Shh) planted ectopically into the anterior side of developing chick limbs leads to the development of a mirror hand.^{28,30} Inappropriate expression of Shh in naturally occurring mouse mutants

has led to the formation of extra ulnar digits.²⁸ Mice deficient in Bone-Morphogenic Protein (BMP)-7 also develop polydactyly.²⁸ It is well known that BMP-7 is expressed at high levels within the interdigital mesenchyme, which undergoes apoptosis in the process of formation of the digital rays.²⁸ Lack of BMP-7 allows for survival of these interdigital cells, which could allow for the formation of extra digits. Another possible factor is the transcription factor, Gli-3, which is important in Shh signalling.²⁸ Mutation or lack of Gli-3 causes polydactyly in both humans and mice, and specifically Pallister-Hall syndrome in humans.³¹ Gli-3 has been shown to be induced by BMP, suggesting that these two factors play a common role in the formation of the digits and in mediating the suppression of polydactyly.²⁸

A family of transcription factors called HOX or Homeobox genes is also important in limb development. Mutations in the specific location of the Hox D13 gene have recently been shown to be the cause of several types of human synpolydactyly.^{1,28}

Study

Materials and Methods

One hundred and thirty six patients with Type B ulnar polydactyly, seen at the Congenital Hand Unit at the Red Cross Children's Hospital, were prospectively reviewed. They were seen over a two-year period from September 2005 to September 2007 and treated using a vascular clip. Their average age was five months (Range: two months – eight years). There were 73 males and 63 females.

Eighty-three patients had bilateral involvement, 50 patients had involvement of the left hand only, three of the right hand, with a total of **219** digits treated. Thirty-three of 62 patients questioned gave a positive family history of Type B ulnar polydactyly.

None of the patients had pre-procedure X-rays as bony articulation with the metacarpal was excluded clinically, prior to the procedure.

Written informed consent was obtained for all patients.

The procedure was performed in the outpatient department at the time of initial presentation. In the young child, local anaesthetic cream (*Emla® 5% cream, lignocaine/prilocaine*) was applied to the base of the rudimentary digit; otherwise, a local anaesthetic injection was given in the older child (*Lignocaine Hydrochloride-Fresenius 2%*). The skin was prepared in a sterile manner and a large Ligaclip® (*Ethicon, Somerville, New Jersey, U.S.A*) vascular clip was used in all cases, irrespective of the age of the child or size of the extra digit. The vascular clip (Figure 1) was applied to the base of the digit, flush with the ulnar border of the hand (Figure 2).

The extra digit was then excised with a No. 23 blade (Figure 3).

No dressing was applied.

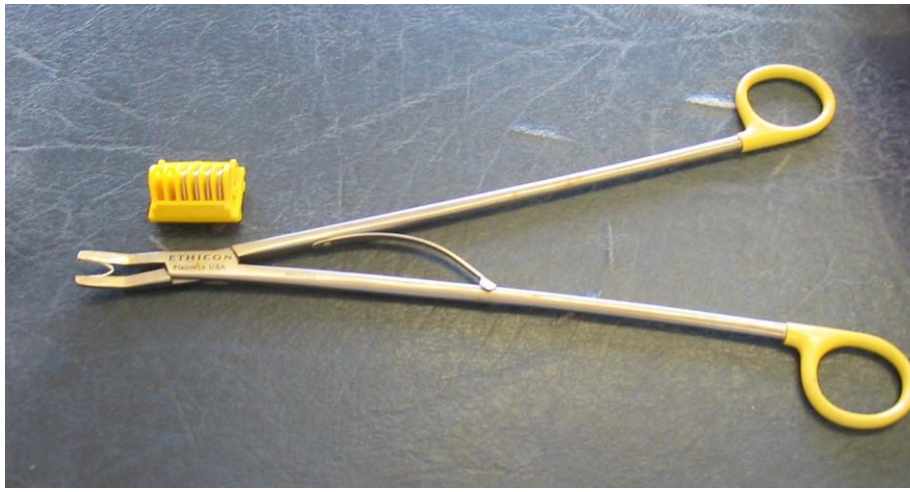


Figure 1: Standard large vascular ligacip® used



Figure 2: Vascular clip application.

Materials and Methods



Figure 3: The clip applied at the base of the digit, flush with the radial border of the hand.

Results

Follow-up was done at one week and again at 3 months.

In 134 of 136 patients, no complications of bleeding, premature loosening of the clip, sepsis or residual nubbin were noted at either visit. Because the extra digit was excised, there were no complications of failure of the digit to autoamputate or venous engorgement. The stump had necrosed at a week, with necrosis occurring on the radial aspect of the clip. At the three month visit, there was no residual nubbin, nor a tender or aesthetically unappealing scar (Figure 4).



Figure 4: Result at 3 months. Necrosis has occurred on the radial border of the clip, leaving no residual nubbin.

In 2/136 patients, however, the clip slipped off at the time of its application, immediately after excision of the extra digit, despite using a large clip. The defect had to be sutured in the outpatient department.

We noted that in both these cases, the base of the extra digit was wider than 4 mm. We recorded the width of the base in all subsequent cases (n = 113).

Even though the literature describes Type B polydactyly as being typically rudimentary and pedunculated, it was noted in this study that this was in fact not always so. Instead, only 55 % of patients had the classic hourglass morphology and 45 % had a much broader or sessile neck. Of the latter “sessile” group, 96 % had a base between 2 – 4 mm, regardless of patient age. Only 4 % had a base wider than 4 mm. When the digit had a base wider than 4 mm, it was technically more difficult to apply the vascular clip, and patients tended to experience discomfort during the procedure. The only two complications also occurred within this sub-group.

Using this data, Type B was further classified into two sub-types according to the width of its base, and not according to its morphology. With this, the unit’s treatment protocol changing accordingly:

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Type 1: Pedunculated type: Base 1 – 4 mm (96 %)

Type 2: Sessile type: Base wider than 4 mm (4 %)

All extra digits with a base wider than 4 mm, that is, U.C.T. Type 2, were subsequently excluded from the study. Instead they were referred for formal excision at one year of age (as for Temtamy and McKusick Type A polydactyly). The vascular clip technique was subsequently only applied to those digits that had a base less than or equal to 4 mm. i.e. U.C.T. Type 1, which formed the majority of the patient cohort (96 %).

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Cost:

The costs of the vascular clip group were compared to that of digits undergoing formal excision over an equivalent period of two years. Patients underwent formal excision as a day case procedure at Red Cross Hospital from July 2004 – July 2006.

There were a total number of 49 digits surgically treated during this time, and the average age at the time of formal excision was 1.5 years (Range: nine months – five years). The average theatre time was 25 minutes (Range 10 – 30 minutes), inclusive of anaesthetic time (the majority of cases were bilateral).

Comparison was made with regard to the cost of the procedure as well as the costs incurred by the patients for the outpatient (OPD) visits (Table 6). Costs for the procedure were calculated at government hospital rates, that is, per procedure, and not for the duration of the procedure or for consumables used.

Table 6: Cost Comparison

	<u>Formal Excision</u>	<u>Vascular Clip</u>
Surgical time per procedure	25 minutes	10 minutes
Surgical cost per procedure	R1182.00	R23.00 (per clip)
Anaesthetic cost per procedure	R163.00	R10.00
Total cost per procedure	R1345.00	R33.00
Average number of OPD visits	5 (minimum)	3 (maximum)
Average cost of OPD visits	R585. 00	R351.00

The total cost of formal excision of 49 digits was R65 905.00 whereas the total cost of an equivalent number of patients who underwent vascular clip ligation was R1 617.00. Thus, the total saving to the hospital over this two-year period for the treatment of 49 digits amounted to R64 288.00. This also translates into a saving of 2 850 theatre minutes. During this period, 219 digits were treated with the vascular clip compared to only 49 digits treated with formal excision.

The patient was billed R117.00 for each outpatient visit / consultation. The average saving to the patient / patient family was thus R234.00. This calculation only included the savings due to the reduced number of outpatient visits. Other unforeseen costs incurred by the family, such as transport costs or time taken off work, school etc was not incorporated when calculating the total cost to the patient.

No comparison was made to the cost of suture ligation, as nurseries keep no record of patients undergoing suture ligation.

Discussion

Traditional treatment of ulnar polydactyly is suture ligation in the immediate post-natal period^{1,3,23} or failing this, formal excision under general anaesthetic when the child is one year of age. However there is some controversy regarding the treatment of Temtamy and McKusick Type B.⁵

Flatt,⁷ Kozin,¹¹ Watson and Hennrikus⁵ and Simmons³² recommended suture ligation for all; Tachdjian³³ recommended division with a blade and electrocautery of the stump. Heras et. al⁸, as well as Leber et. al⁹ recommended surgical excision for all supernumerary digits to avoid post-traumatic neuromas associated with suture ligation; whereas Jobe²⁰ suggested formal excision and skin closure as the only form of treatment, discouraging ligation because of reports of fatal haemorrhage following suture ligation.

Dodd et. al.³⁴ conducted a survey to review current practice in managing supernumerary digits in the United Kingdom, when it was noted that there was uncertainty amongst paediatricians about the management of this common anomaly. The survey involved both paediatricians and hand surgeons and was done via a questionnaire with three attached clinical photographs of Type B polydactyly of various sizes. Case 1 was of a wart-like nubbin, Case 2, a pedunculated digit, and Case 3 a well-formed digit with a wide base, similar in appearance to a Type A duplication. The decision was unanimous regarding the management of Case 3. However, there was no consensus amongst paediatricians and neonatologists regarding the recommended practice for the other two cases, particularly of Case 2, the most commonly occurring morphology. The majority of paediatricians (79 %) indicated that they would not treat Cases 1 and 2 themselves, but instead advocated specialist referral for this common group. Referral was three times more common if the specialist service

was available on-site. There was also no consensus regarding the timing of intervention, and only one-third of both paediatricians and hand surgeons thought it was appropriate that Case 2 be treated within one month of birth. Thus, there is marked variation in management, even amongst specialists, about all but the most complex forms of a simple anomaly such as Type B polydactyly.

In South Africa, the majority of patients with Type B ulnar polydactyly are treated with suture ligation in the neonatal period. When these are missed during this early period, they are treated by formal excision under general anaesthetic (G.A.) once the child reaches one year of age. These treatment interventions however, are associated with many complications. The highest complication rates are associated with suture ligation.³ These complications will be discussed further.

Suture Ligation

Many regard suture ligation as the standard of care for Type B polydactyly and is advocated in many paediatric and neonatology textbooks.⁹ It is often performed in the neonatal or paediatric unit in the immediate post-natal period.^{5,9,23} Unfortunately, suture ligation is associated with complications such as bleeding, a report of fatal haemorrhage⁷, failure of the digit to autoamputate, venous congestion, gangrene³⁶, infection,^{1,3,5,35} sensitive residual nubbins³ and traumatic neuromas.^{8,9} The task of ligation often falls to the most junior or inexperienced personnel in the neonatal unit, with poor surgical knot tying technique. Added to this is the technically demanding task of applying a suture to a rudimentary digit in a neonate who refuses to remain still. As a result, the suture may not be placed right at the base of the digit, flush with the ulnar border of the hand. Instead, the suture ends up being tied in the middle of the digit. Thus, when the rudimentary digit eventually autoamputates, the patient is left with a residual nubbin which is often sensitive^{1,3} (Figure 5).



Figure 5: Residual nubbins which are often sensitive

Alternatively, the suture is poorly tied, with subsequent knot slippage, leading to venous congestion of the stump (Figure 6).



Figure 6: Venous congestion of the extra digit

This may be complicated by secondary infection, or failure of the gangrenous stump to auto-amputate.^{3,5,35} Watson and Hennrikus⁵ prospectively reviewed 28 digits treated with suture ligation, followed up for an average period of 20 months. Forty-three percent of patients had a residual bump ranging from 2 - 6 mm. None had a cosmetically appealing result, and only 57 % had what he referred to as a “small scar that was barely noticeable”.⁵ In both this series and that by Flatt,⁷ several patients required formal excision as a secondary procedure, as the necrotic digit had failed to auto-amputate after a month.^{1,5,7} Upton³⁶ reported gangrene in a digit that failed to autoamputate, as well as several cases of infection. Frieden et. al.³⁵ also reported a complication of secondary bacterial infection following suture ligation. Rayan and Frey³ reviewed 148 patients with both Type A and B polydactyly, and reported a higher complication rate with suture ligation than with surgical treatment. In the suture ligation

group, 24 % developed wound infection. Seventeen patients were left with residual nubbins. Two patients in this series required re-operation for sensitive residual nubbins, and one for continued bleeding after the remnant had sloughed.

Several authors also postulate that these sensitive nubbins are in fact traumatic neuromas because of suture ligation in infancy.^{8,9,23} Heras et al.⁸ reported one case where the digit “reformed” a year after suture ligation. Histology following surgical excision confirmed a traumatic neuroma. Leber et. al.⁹ reported 3 cases of patients who presented with nubbins on the ulnar border of the hand, requesting excision for sensitivity, as well as for cosmetic reasons. Their ages ranged from 1 year to 13 years. All patients were treated with elliptical surgical excision and proximal division of the digital accessory nerve. Histological studies after excision of the supernumerary digit confirmed neural proliferation consistent with a traumatic amputation neuroma.⁹ None had any recurrence of the traumatic neuroma or a cosmetically unappealing scar after a follow-up of 2 - 6 years. Interestingly, on questioning the parents, all had had ulnar polydactyly themselves, treated with suture ligation when they were neonates. All still experienced hypersensitivity, occasional bleeding and ulceration, but had never sought further medical treatment. For these reasons, the authors felt that suture ligation results in neuromas that are symptomatic and “yield a poor surgical result”.⁹ Thus, they advocate surgical excision as the only acceptable form of treatment for rudimentary ulnar digits, in order to properly address the accessory digital nerve and avoid the complication of traumatic neuroma formation and the sensitive residual nubbin.^{8,9}

Formal excision

Formal excision is recommended when the digit was missed or not treated with suture ligation in the neonatal period.²⁰ This is routinely only done at six months to one year of age,²⁰ to minimise the risks of a general anaesthetic. The digit is removed through an elliptical incision. Anomalous tendons are removed, the digital nerves incised and the digital arteries coagulated. If the ablated digit contains the abductor digiti minimi (ADM), the tendon and muscle need to be transferred to the adjacent digit to restore abduction.¹¹ The ulnar collateral ligament may need to be reconstructed if the extra digit is attached at the level of the metacarpophalangeal joint.³⁷

Excision carries with it the inherent risks of an anaesthetic as well as occasionally leaving a sensitive scar on the ulna border of the hand. Certainly in Type A polydactyly, but also in some cases of Type B, where the digit is more well developed than the classically described nubbin, other complications of excision present themselves. Injury to the ulnar collateral ligament, extensor tendon imbalance, inadequate resection of the proximal phalangeal head leading to an unsightly bump (particularly when there is a bifid metacarpal articulation), or retained cartilage and a stiff metacarpophalangeal joint are all well-recognised complications of surgical excision.^{10,20}

Formal excision demands the availability of health facilities which can offer paediatric anaesthesia and recovery services, as well as a dedicated children's ward and staff. It also requires numerous visits by the patient: the initial clinic visit, the repeat visit once the child has reached one year of age, the day of surgery and usually two post-operative visits. Often these children have intercurrent illness (usually upper respiratory tract infections) on the day of surgery, thus they have to be cancelled and rebooked, incurring further expenses. In our society, and considering the

disadvantaged population that Red Cross Children's Hospital serves, the majority of our patient population can ill afford this expense.

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Vascular clip ligation

This study confirms the extremely common nature of ulnar polydactyly in South Africa. The reported incidence in South Africa is 1 in 100 live births, making it possible to treat 136 patients in less than three years. Blauth and Olason¹⁵ treated 134 patients over a 30-year period; Rayan and Frey³ treated 148 patients over 10 years, whereas Watson and Hennrikus⁵ only managed to treat 28 digits over a three-year period.

In this cohort, the male to female ratio was 1.5:1. The majority of the patients had bilateral involvement (61 %) and when unilateral, had a definite preponderance for the left side (94 %), which is keeping with the literature, where the left side is involved in approximately 70% of unilateral cases.^{10, 14, 22} Of 136 patients, only three patients (6%) had involvement of the right hand. The reason for this has never been explored or explained.

In this study, 53% of patients had a family member who also had ulnar polydactyly, either a parent or grandparent. The reported incidence of a positive family history of ulnar polydactyly varies, but is anywhere from 11- 33%, up to 90%.³⁸

The majority of the patients (81 %) were of African descent (n=110), the rest were either Caucasian or of Mixed race. None of the study patients had any known associated syndromes, however none of the Caucasian patients were referred to a geneticist to confirm this assumption.

Despite Temtamy and McKusick's opinion that there are only two types of ulnar polydactyly, we have recognised from our experience, and agree with Rayan and Frey,³ as well as with Al-Qattan,¹⁴ that there is in fact a spectrum of presentation of the rudimentary digit within the Temtamy and McKusick Type B group. Ligation, therefore, whether it be by suture or vascular clip, is not appropriate in all instances.

Not all digits seen at the Red Cross Children's Hospital Congenital Hand Clinic were rudimentary and pedunculated as classically described (Figure 7).



Figure 7: The classic Type B pedunculated digit (U.C.T. Type 1)

Nearly half (45 %) had a much broader or sessile neck (Figure 8), as opposed to the classic hourglass morphology, despite all being classified as “rudimentary” (i.e. cutaneous appendage, no osseous component, no articulation with the metacarpal^{4,13}).



Figure 8: The U.C.T. Type 2 Sessile type

This is in keeping with a similar finding by Al-Qattan et. al.¹⁴ who described a range of pedicle width within Temtamy and McKusick Type B or Rayan and Frey Type II group (See Table 3). This led them to modify Rayan and Frey's classification and sub-classify Type II into a Type IIA and IIB, where the pedicle width is less than or more than 3 mm, respectively. However we found that when the base was wider than 4 mm, (and not 3 mm as in their study), the application of the vascular clip was technically more difficult. The only two complications in the Red Cross Hospital study occurred within this group and patients tended to experience discomfort during the procedure despite local anaesthetic use, presumably due to the greater amount of tissue being crushed by the clip.

As a result, the unit's practice changed and Type B polydactyly was subsequently classified according to the width of the base of the extra digit. Type B polydactyly was sub-classified into two groups to determine the management of the extra digit (Table 7).

Table 7: Red Cross Hospital Congenital Hand Unit Classification

<u>University of Cape Town</u>	<u>Morphology</u>	<u>Management</u>
Type 1	Pedunculated type: Base \leq to 4 mm	Vascular clip ligation
Type 2	Sessile type Base $>$ 4 mm	Refer for Formal excision

Type 1 is the most common presentation (96 %) and should be treated by vascular clip ligation at initial presentation, regardless of age of presentation. Type 2 is rare (4 %) and treatment should instead be as for Type A polydactyly; that is, early referral to a dedicated specialist hand unit for formal excision under general anaesthesia when the child is one year of age, to avoid any potential complications and patient discomfort.

For various reasons which will be discussed, vascular clip ligation, in our setting, makes an excellent and cost-effective alternative to both suture ligation and surgical excision in the treatment of Type B polydactyly.

Type B polydactyly can be treated as an outpatient procedure in non-specialist centres. These rudimentary digits can be treated at the time of initial presentation to the clinic, requiring only a brief procedure in an often-busy clinic. In this regard, it is advantageous when compared to surgical excision as it obviates the need for repeated visits until the child is old enough to tolerate a general anaesthetic and avoids the potential complications of both an anaesthetic and surgical procedure in the infant.

One hundred and thirty six patients were treated with vascular clip ligation during this study with only two recorded complications.

In 219 digits treated, none were seen with any of the early complications associated with suture ligation, such as bleeding, pain, venous engorgement, sepsis of the stump or a residual nubbin. Because there were no complications noted at the one week or at the three-month visit, and all digits had healed with an aesthetically appealing scar, none of the patients were asked to return for long-term follow-up.

During the study period, no patients returned with “recurrence” of the digit or with traumatic neuromas as reported by Heras et. al.⁸ However, the study patients were only followed-up for 3 months and it is well documented that these neuromas can take up to 1-2 years to develop.⁹

The vascular clip is technically easier to apply than sutures, due to better control during placement of the clip. It therefore does not require the necessary skill needed for placing a proper surgical knot, making it more suitable for use by non-surgical or more inexperienced medical practitioners. As the application of the clip can be better controlled than when placing a suture, it is easier to apply the clip to the base of the extra digit, flush with the ulnar border of the hand. Thus, when the rudimentary digit sloughs and necroses, it does so on the radial side of the clip, leaving no residual nubbin.

It is accepted that the low complication rate is in part due to the fact that the procedure was performed by dedicated hand surgeons. However, the technique of clip application is specific but uncomplicated. Therefore it is recommended that the medical and nursing staff involved in the nursery would need to undergo simple training in the correct technique of clip application, to avoid any potential complications such as premature loosening of the clip, or those complications associated with suture ligation listed above. To address the issue, a treatment algorithm

has been included (See Addendum 1) which could be made available to all staff treating ulnar polydactyly.

The clip need not exclusively be used to treat neonates, but can also be used to treat children and adults, though the oldest patient treated in this study was eight years of age. There should be no contraindication to using the clip in an older age group, provided the base of the extra digit is ≤ 4 mm.

This procedure also represents a major cost-effective alternative, with significant savings per patient treated. This saving translates to both the hospital and the patients' families. The total saving to the hospital over the study period for the treatment of 49 digits amounted to R64 288.00. This was when compared to 49 patients undergoing formal excision under general anaesthesia. When one extrapolates this amount to the treatment of 219 digits, the total saving to the hospital would be approximately R 287 328.00.

Because the vascular clip can be applied at the first outpatient visit, the family needs to attend the hospital for an average number of three visits. This is opposed to waiting until the child is old enough to have a formal excision under G.A., where a minimum of five visits is required.

The total average saving to the patient / patient family was R234.00. This did not include the other unforeseen costs incurred by the family, such as transport costs, time taken off work and school and alternate arrangements that needed to be made to enable parents to escort the child to the hospital.

In government hospitals, patients are charged per procedure. In a private hospital, the cost savings would be more significant, as patients are charged not only per procedure, but also for theatre time and all consumables used. This only includes hospital costs; it does not include the surgeon and anaesthetist's fee. The saving to the patient would be substantial when using vascular clip ligation, as the procedure can be

performed in the doctor's consulting rooms, under local anaesthesia, at a fraction of the total cost.

Because Type B ulnar polydactyly is extremely common in South Africa, a large number of children are seen annually at Red Cross Children's Hospital Congenital Hand Clinic with this anomaly. The policy at the unit was previously for all patients with supernumerary digits that had been missed in the neonatal period, to have formal excision under general anaesthesia once the child reached the age of 1 year, as suture ligation was no longer a viable option due to the high complication and failure rate. Unfortunately, this required theatre time and staff and the waiting list for formal excision was extremely long. These cases occupied theatre lists to the exclusion of all else. Since the introduction of the vascular clip, an excess of 3000 theatre minutes have been saved. More than double the number of patients have been treated with a vascular clip compared to that by formal excision within the same time period.

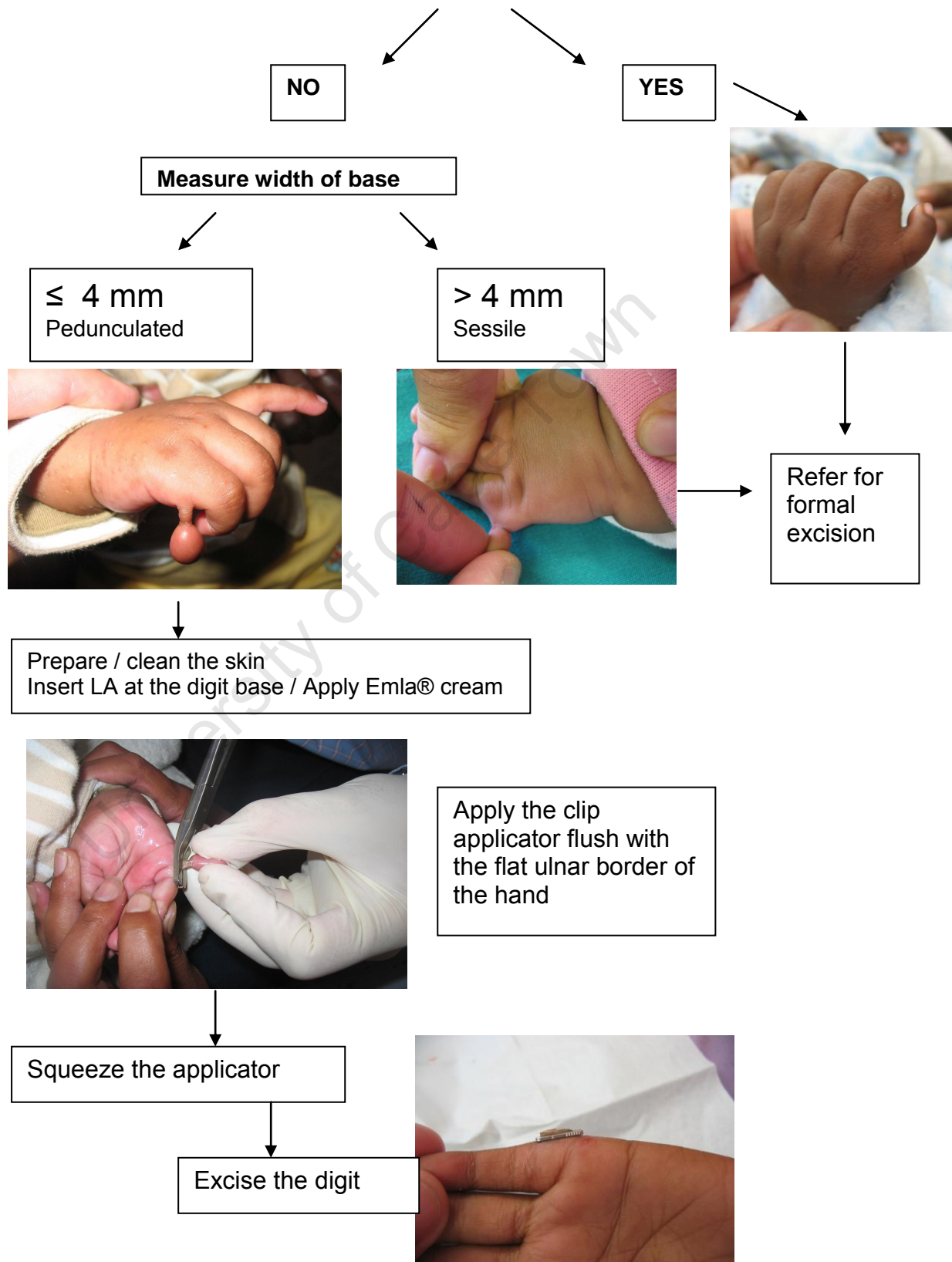
It would be advantageous if vascular clips became available in all settings where these extra digits are treated, especially in obstetric and neonatal units. Considering the lack of consensus amongst paediatricians regarding the treatment of Type B, it would be ideal if the simple treatment algorithm mentioned below (Addendum 1) could be included in the curriculum of the paediatric registrar and that they gain exposure during their registrar training to the simple technique of applying the vascular clip.

The recommendations of this study are:

1. That vascular clips and applicators be made available in all obstetric and neonatal units.
2. That all paediatric registrars as well as nursing staff, dealing with a paediatric patient population, receive the necessary exposure and training in the application of the vascular clip.
3. A simple treatment algorithm is followed.

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Treatment algorithm: Ulnar polydactyly
Is there bony articulation with the metacarpal?



Conclusion

Polydactyly is one of the most common congenital hand anomalies and almost every hand surgeon will encounter this problem at some point. The surgeon therefore needs to appreciate the anatomical variety, the systemic associations as well as the reconstructive options.¹

University of Cape Town Type 1 is the most common presentation (96%) and should be treated by vascular clip ligation at initial presentation, regardless of age of presentation.

University of Cape Town Type 2 is rare (4%) and treatment should instead be as for Type A polydactyly; that is, early referral to a dedicated specialist hand unit for formal excision under general anaesthesia when the child is one year of age, to avoid any potential complications and patient discomfort.

Vascular clip ligation should become the standard of care for Type B ulna polydactyly, as it is a better alternative to conventional forms of treatment in terms of its ease of application, better cosmetic result and low complication rate. It is certainly more cost-effective to the hospital, the attending doctor but probably most importantly, to the patient.

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